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Case Report

Congenital coronary artery fistula presenting later in life

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Summary A 53-year-old male presented to our tertiary medical center with complaints of dyspnea and exertional chest pain with mild left ventricular dysfunction and right ventricular enlargement on echocardiography. Cardiac catheterization showed a congenital right coronary artery fistula communicating with the right sided chambers. Using contrast enhanced multi-detector computed tomography scan, the fistula was clearly draining into the coronary sinus. We describe briefly the etiology of coronary artery fistula, its clinical presentation, and the common tests used to confirm diagnosis. We further discuss the types of treatment modalities that are currently available.

Introduction

Congenital coronary artery fistulae are rarely seen in our daily practice. They comprise less than 1% of all congenital heart defects [2]. Symptoms occur in childhood or early adulthood depending on the type of fistula, size, and shunt severity [1]. Congenital coronary artery fistulae are usually found incidentally on coronary angiograms. Coronary computed tomography angiogram is a helpful non-invasive method to diagnose coronary artery fistula as it can delineate the nature of the fistula, number, origin and insertion sites. Patients can be managed conservatively or may require fistula closure with increased shunt severity (Qp/Qs >1.5) [1]. This is done surgically or using percutaneous transcatheter closure devices based on type and nature of the fistula [5]. We describe below a patient who presented in late adulthood with a right coronary artery fistula to the coronary sinus and significant left to right shunting. We review briefly the etiology of coronary artery fistulae, clinical presentation, diagnostic tests used to establish diagnosis and current treatment modalities.

Case report

A 53-year-old male with chronic persistent atrial fibrillation and hypertension presented with several year history of

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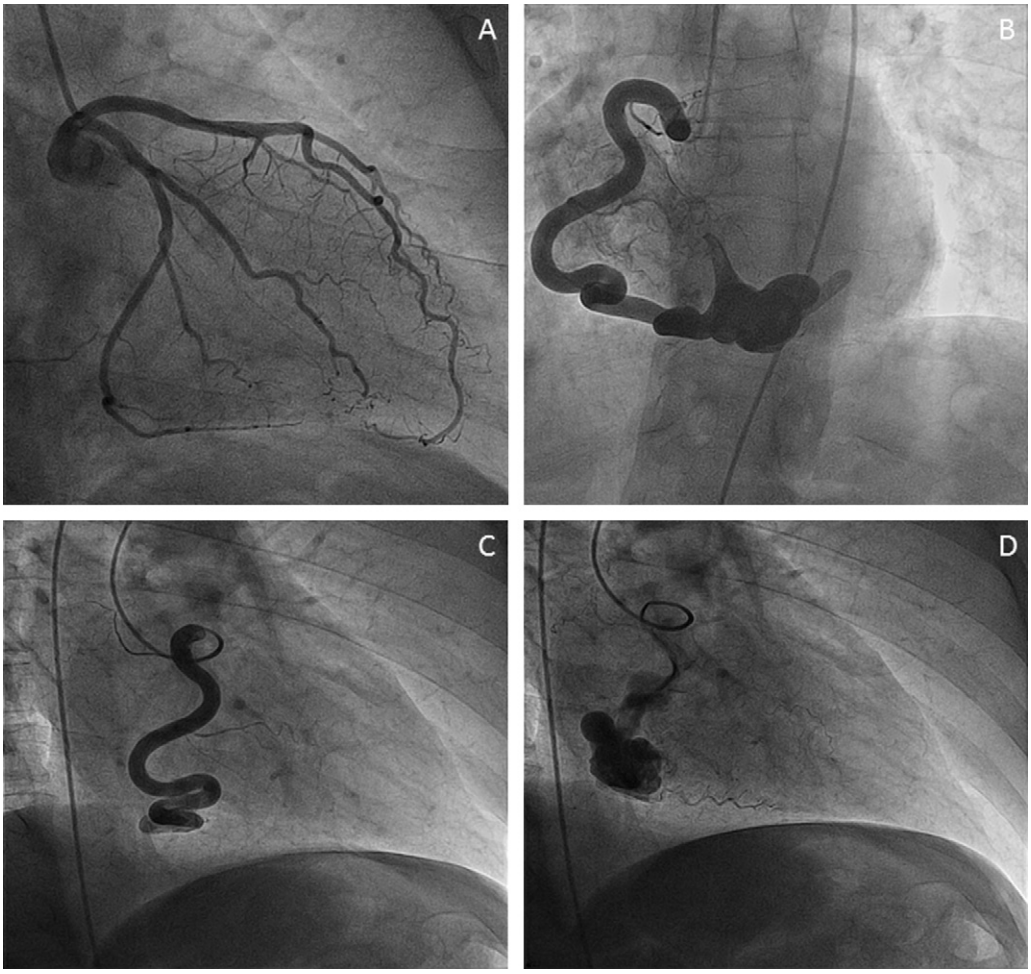

Figure 1 Coronary angiogram showing: (A) RAO caudal view of the left coronary artery with normal LAD and LCX arteries. (B) AP view of the RCA with a markedly dilated and tortuous RCA with apparent fistula draining into right-sided chambers. (C) RAO view showing a dilated RCA with increased tortuosity distally. (D) High-density contrast is seen in the right atrium consistent with a RCA fistula communicating with the right sided chambers. AP, anteroposterior; CAF, coronary artery fistula; LAD, left anterior descending; LCX, left circumflex; RAO, right anterior oblique; RCA, right coronary artery.

worsening dyspnea, fatigue, and exertional chest pain. Physical exam, chest X-ray, and serial cardiac enzymes were all unremarkable. Electrocardiography (ECG) showed atrial fibrillation rate controlled. Transthoracic echocardiography showed mildly reduced left ventricular function with basal inferoposterior wall hypokinesis, mild biatrial and right ventricular enlargement. The patient underwent conventional coronary angiography that showed normal left coronaries and a massively dilated right coronary artery (RCA) communicating with the right atrium (RA) (Fig. 1). Right cardiac catheterization confirmed the above findings with step up in oxygen saturation from 61% (RA) to 76% (right ventricle and pulmonary artery). A left to right shunt (Qp/Qs 1.4) was present; mean pulmonary artery pressure and wedge pressure were 20 mmHg and 15 mmHg respectively. As distal drainage site was not well identified, a dedicated contrast-enhanced ECG-gated multidetector computed tomogram (GE 64 multidetector CT scanner; GE Healthcare, Milwaukee, WI, USA) was performed that clearly demonstrated a tortuous and markedly dilated RCA, giving rise to a large fistula that drained into the coronary sinus (Fig. 2). Due to extreme vessel tortuosity and difficulty in cannulating the distal fistula, the patient was not a candidate for percutaneous transcatheter closure and was referred for surgical ligation of the fistula by cardiothoracic surgery. The patient requested postponing his surgery for at least a year and to be managed conservatively.

Discussion

This patient's coronary artery fistula (CAF) is a rare congenital form of coronary artery anomalies previously described in the literature [1] with an incidence of 0.2—0.47% of all congenital cardiac defects [2]. CAF can be also acquired secondary to trauma that is either iatrogenic or accidental [3]. Clinical presentation varies according to etiology, age, and duration of shunt. In congenital CAF, around 20% of patients are symptomatic in the first two decades that later increases to 60% after the age of twenty years. Neonates and infants may present with heart failure with left to right shunting or cyanotic heart disease depending on shunt type.

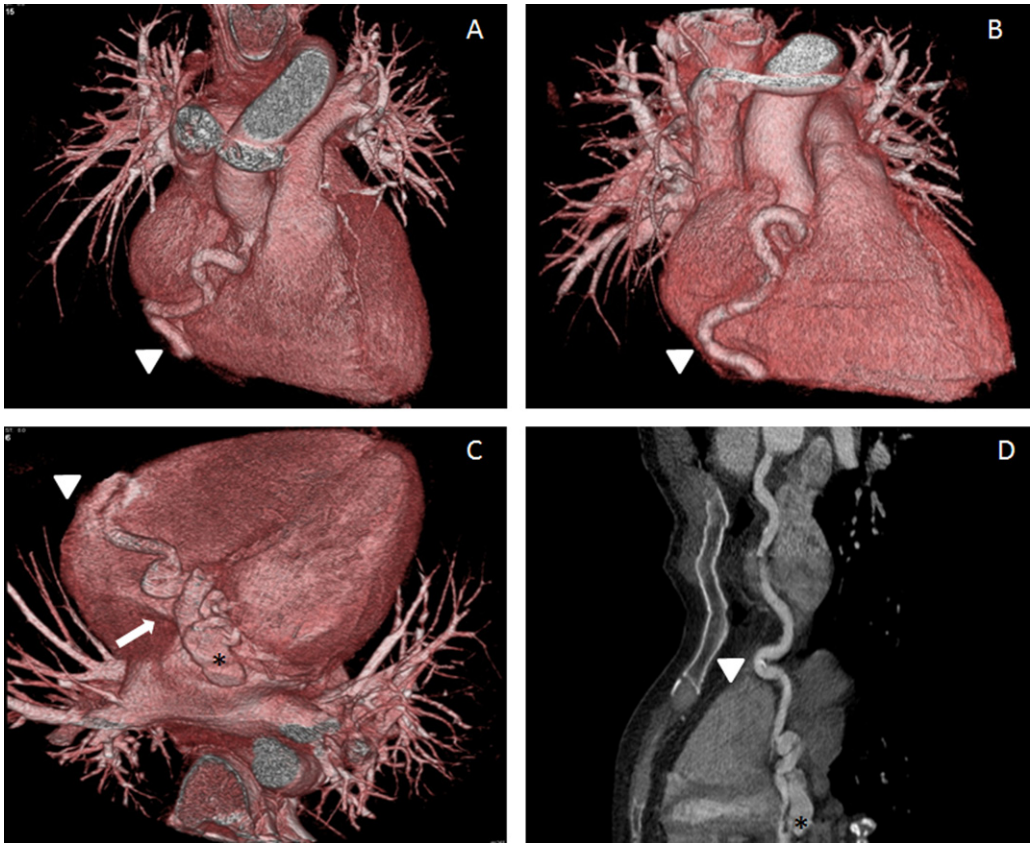

Figure 2 (A—C) Volume rendered images from coronary computed tomography angiogram showing a tortuous and largely dilated RCA (arrow heads) most prominent on the inferior border of the heart. The asterisk points out the fistula communication between the RCA and coronary sinus. The arrow points out the entry of the sinus into the right atrium. The right atrium appears mildly dilated. (D) Curved planar reformatted image showing a dilated tortuous RCA draining distally into the coronary sinus. RCA, right coronary artery.

Older children experience exercise-induced angina. Adults present with shortness of breath, fatigue, exercise intolerance that worsen with shunt augmentation, and progressive enlargement of the fistula [1]. On physical exam, a mild mid diastolic murmur is audible when the fistula drains to the right ventricle. Machinery like murmur at the left upper sternal border is heard when the fistula drains into the pulmonary artery, mimicking a persistent ductus arteriosus [1]. CAF may eventually lead to premature atherosclerosis secondary to shear-induced intimal damage caused by the turbulent blood flow, angina, myocardial infarction, saccular aneurysms, arrhythmias, and/or infective endocarditis. Large CAFs cause angina due to coronary steal phenomena. Distal intracoronary diastolic pressure is reduced and increased vessel size may lead to myocardial ischemia. Large fistulae dilate over time increasing the risk of thrombosis, endocarditis, or rupture.

Chest radiography and ECG findings are usually normal except in large fistulae where there may be cardiomegaly and signs of volume overload. Echocardiography with color flow Doppler is useful in the assessment of shunt physiology. Coronary angiography with a right heart catheterization is the diagnostic test of choice to evaluate the course of the fistula and its anatomy and to calculate shunt hemodynamics (Qp/Qs). In some cases, it is difficult to locate the origin of the fistula and its insertion site. To better delineate the cardiac anatomy, the number of fistulae, size, origin, and drainage sites, a 64 multidetector computed tomography is extremely helpful with good spatial and temporal resolution. Many scholars consider it as the test of choice to help guide therapy as in the case we presented above.

The treatment goal is to obliterate the fistula while preserving normal coronary blood flow. The risk of conservative treatment should be balanced with the risk of procedure used to occlude the fistula. In general fistulae with Qp/Qs ratio > 1.5 require closure. Treatment of acquired CAF depends whether it is accidental or iatrogenic. Accidental traumatic CAFs result in long term complications, therefore immediate repair is recommended. Iatrogenic traumatic fistulae on the other hand have a benign clinical course and most heal spontaneously with 75% of the cases resolving on follow up angiography. In congenital CAF, repair is recommended if the patient is symptomatic in childhood or develops complications in adulthood or when Qp/Qs > 1.5 regardless of symptoms. Repair of smaller shunts is still controversial and two opinions are currently present. One favors surgical closure early in childhood due to lower postoperative mortality and a high success rate in this patient population [1]. Scholars argue that even if the patient is asymptomatic, serious complications such as myocardial

ischemia, chronic heart failure, endocarditis, or aneurysmal dilatation can be prevented [1]. Others advocate that tiny newly diagnosed CAF be treated conservatively with follow up surveillance examinations [4]. The incidence of cardiac complications is low and there is always a possibility of spontaneous closure.

Two treatment modalities are available for fistulae closure: surgical vs. percutaneous transcatheter closure (TCC). Surgical repair includes: (1) external ligation via direct visualization of the fistula with no need for cardiopulmonary bypass; or (2) closure from within the recipient chamber which is a more extensive surgery and requires cardiopulmonary bypass [1]. TCC is a less invasive method, most effective with proximal fistulas that have single origins and draining sites [5]. Several embolic materials are available to close the fistula including coils, detachable balloons, double-umbrella, and particles [5]. Coil embolization is most frequently used with successful results when used in the absence of multiple fistulas, a single narrowing draining site, absence of large branch vessel, and safe accessibility to the coronary artery supplying the fistula. Complications with TCC include proximal or distal coil migration, thrombosis of parent epicardial vessel, distal undersizing of the coil resulting in a high shunt flow, coronary artery spasm, fistula dissection, dysarrhythmias, and infective endocarditis [5]. Patients are placed on antiplatelet therapy and in some cases on short-term anticoagulation and antibiotics. Few case reports and case series have shown comparable outcomes with transcatheter closure and surgery without the associated morbidities of cardiopulmonary bypass and/or sternotomy in the surgical group [5].

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